



General

Guideline Title

Pheochromocytoma and paraganglioma: an Endocrine Society clinical practice guideline.

Bibliographic Source(s)

Lenders JW, Duh QY, Eisenhofer G, Gimenez-Roqueplo AP, Grebe SK, Murad MH, Naruse M, Pacak K, Young WF Jr, Endocrine Society. Pheochromocytoma and paraganglioma: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab. 2014 Jun;99(6):1915-42. [264 references] PubMed

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Recommendations

Major Recommendations

Definitions for the quality of the evidence (+OOO, ++OO, +++O, and +++++); the strength of the recommendation (1 or 2); and the difference between a "recommendation" and a "suggestion" are provided at the end of the "Major Recommendations" field.

Biochemical Testing for Diagnosis of Pheochromocytoma and Paraganglioma (PPGL)

The Task Force recommends that initial biochemical testing for PPGLs should include measurements of plasma free metanephrines or urinary fractionated metanephrines. (1|+++++)

The Task Force suggests using liquid chromatography with mass spectrometric or electrochemical detection methods rather than other laboratory methods to establish a biochemical diagnosis of PPGL. (2|+++OO)

For measurements of plasma metanephrines, the Task Force suggests drawing blood with the patient in the supine position and use of reference intervals established in the same position. (2|+++OO)

The Task Force recommends that all patients with positive test results should receive appropriate follow-up according to the extent of increased values and clinical presentation. (1|++OO)

Imaging Studies

The Task Force recommends that imaging studies to locate PPGL should be initiated once there is clear biochemical evidence of a PPGL.

(1|++OO)

The Task Force suggests computed tomography (CT) rather than magnetic resonance imaging (MRI) as the first-choice imaging modality because of its excellent spatial resolution for thorax, abdomen, and pelvis. (2|+++O)

The Task Force recommends MRI in patients with metastatic PPGL, for detection of skull base and neck paragangliomas, in patients with surgical clips that cause artifacts when using CT, in patients with an allergy to CT contrast, and in patients in whom radiation exposure should be limited (children, pregnant women, patients with known germline mutations, and those with recent excessive radiation exposure). (1|++++O)

The Task Force suggests the use of ¹²³I-metaiodobenzylguanidine (MIBG) scintigraphy as a functional imaging modality in patients with metastatic PPGL detected by other imaging modalities when radiotherapy using ¹³¹I-MIBG is planned, and occasionally in some patients with an increased risk for metastatic disease due to large size of the primary tumor or to extra-adrenal, multifocal (except skull base and neck PPGLs), or recurrent disease. (2|+OOO)

The Task Force suggests the use of ¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) positron emission tomography (PET)/CT scanning in patients with metastatic disease. ¹⁸F-FDGPET/CT is the preferred imaging modality over ¹²³I-MIBG scintigraphy in patients with known metastatic PPGL. (2|++++O)

Genetic Testing

The Task Force recommends that all patients with PPGLs should be engaged in shared decision making for genetic testing. (1|++++O)

The Task Force recommends the use of a clinical feature-driven diagnostic algorithm to establish the priorities for specific genetic testing in PPGL patients with suspected germline mutations. (1|+++O)

The Task Force suggests that patients with paraganglioma undergo testing of succinate dehydrogenase (SDH) mutations and that patients with metastatic disease undergo testing for *SDHB* mutations. (2|++++O)

The Task Force recommends that genetic testing for PPGL be delivered within the framework of health care. Specifically, pretest and post-test counseling should be available. All tests for PPGL genetic testing should be performed by accredited laboratories. (Ungraded recommendation)

Perioperative Medical Management

The Task Force recommends that all patients with a hormonally functional PPGL should undergo preoperative blockade to prevent perioperative cardiovascular complications. (1 \mid ++OO) The Task Force suggests α -adrenergic receptor blockers as the first choice. (2 \mid ++OO)

The Task Force recommends preoperative medical treatment for 7 to 14 days to allow adequate time to normalize blood pressure and heart rate. Treatment should also include a high-sodium diet and fluid intake to reverse catecholamine-induced blood volume contraction preoperatively to prevent severe hypotension after tumor removal. (1|++OO)

The Task Force recommends monitoring blood pressure, heart rate, and blood glucose levels with adjustment of associated therapies in the immediate postoperative period. (1|+++OO)

The Task Force suggests measuring plasma or urine levels of metanephrines on follow-up to diagnose persistent disease. The Task Force suggests lifelong annual biochemical testing to assess for recurrent or metastatic disease. (2|+++OO)

Surgery

The Task Force recommends minimally invasive adrenalectomy (e.g., laparoscopic) for most adrenal pheochromocytomas. (1|++OO) The Task Force recommends open resection for large (e.g., >6 cm) or invasive pheochromocytomas to ensure complete tumor resection, prevent tumor rupture, and avoid local recurrence. (1|+OOO) The Task Force suggests open resection for paragangliomas, but laparoscopic resection can be performed for small, noninvasive paragangliomas in surgically favorable locations. (2|+OOO)

The Task Force suggests partial adrenal ectomy for selected patients, such as those with hereditary pheochromocytoma, with small tumors who have already undergone a contralateral complete adrenal ectomy to spare adrenal cortex to prevent permanent hypocortisolism (2|+OOO)

Personalized Management

In recognition of the distinct genotype-phenotype presentations of hereditary PPGLs, the Task Force recommends a personalized approach to patient management (i.e., biochemical testing, imaging, surgery, and follow-up). (Ungraded recommendation)

The Task Force recommends that patients with PPGLs should be evaluated and treated by multidisciplinary teams at centers with appropriate expertise to ensure favorable outcome. In particular, patients should be referred to such centers should there be pregnancy, metastatic disease, or issues concerning the complexity or difficulty in biochemical diagnosis; localization; performance and interpretation of genetic testing, preoperative preparation; surgical treatment; and follow-up. (Ungraded recommendation)

Definitions:

Quality of Evidence

+OOO Denotes very low quality evidence

++OO Denotes low quality evidence

+++O Denotes moderate quality evidence

++++ Denotes high quality evidence

Strength of Recommendation

- 1 Indicates a strong recommendation and is associated with the phrase "The Task Force recommends."
- 2 Denotes a weak recommendation and is associated with the phrase "The Task Force suggests."

Clinical Algorithm(s)

The following algorithms are provided in the original guideline document:

- Decisional algorithm for genetic testing in patients with a proven PPGL
- Decisional algorithm for functional imaging in patients with proven PPGL

Scope

Disease/Condition(s)

Pheochromocytoma and paraganglioma (PPGL)

Guideline Category

Diagnosis

Evaluation

Management

Screening

Treatment

Clinical Specialty

Endocrinology

Medical Genetics

Nuclear Medicine

Oncology

Pediatric	S
Radiolog	33

Surgery

Intended Users

Advanced Practice Nurses

Physician Assistants

Physicians

Guideline Objective(s)

To formulate clinical practice guidelines for pheochromocytoma and paraganglioma (PPGL)

Target Population

Adults and children with known or suspected pheochromocytoma and paraganglioma (PPGL)

Interventions and Practices Considered

Diagnosis/Evaluation

- 1. Biochemical testing
 - Measurements of plasma free metanephrines or urinary fractionated metanephrines
 - Liquid chromatography with mass spectrometric or electrochemical detection
 - Drawing blood in the supine position for measurements of plasma metanephrines and use of reference intervals established in the same position
 - Follow-up of patients with positive test results
- 2. Imaging studies
 - Computed tomography (CT)
 - Magnetic resonance imaging (MRI)
 - 123I-metaiodobenzylguanidine (MIBG) scintigraphy
 - ¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) positron emission tomography (PET)/CT scanning in patients with metastatic disease.
- 3. Genetic testing
 - Shared decision-making
 - Use of a clinical feature-driven diagnostic algorithm
 - Testing of succinate dehydrogenase (SDH) mutations
 - Pretest and post-test counseling
 - Use of accredited laboratories

Treatment/Management

- 1. Perioperative medical management
 - Preoperative blockade with α-adrenergic receptor blockers
 - Treatment to normalize blood pressure and heart rate
 - High-sodium diet and fluid intake
 - Monitoring blood pressure, heart rate, and blood glucose levels
 - Measuring plasma or urine levels of metanephrines on follow-up
- 2. Surgery
 - Minimally invasive surgery (e.g., laparoscopic adrenalectomy)

- Open resection for selected patients
- Partial adrenalectomy for selected patients
- 3. Personalized management

Major Outcomes Considered

- Changes in the management of index patients
- Number of patients diagnosed biochemically correctly
- Added diagnostic benefit attributable to functional imaging (FI) in localizing primary tumors or metastasis
- Harm resulting from the use of FI
- Proportion of sporadic pheochromocytoma and paraganglioma patients with newly detected germline mutations
- Change in surgical intervention
- Completeness of surgical resection
- Resolution of symptoms

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

The Task Force reviewed primary evidence and commissioned two additional systematic reviews (see the "Availability of Companion Documents" field) to support the guideline.

Testing for Germline Mutations in Sporadic Pheochromocytoma/Paraganglioma: A Systematic Review

Study Eligibility

Studies eligible for inclusion were full publications of observational studies that included patients with sporadic and biochemically proven pheochromocytomas (PCCs) and paragangliomas (PGs) who also underwent germline genetic testing. Diagnoses were considered sporadic when there was (i) a negative family history of PCC/PG, (ii) the absence of syndromic features, (iii) a lack of bilateral disease and (iv) the absence of metastatic disease. The outcomes of interest were the proportion of sporadic pheochromocytoma and paraganglioma (SPP) patients with newly detected germline mutations, the number of family members diagnosed with a new mutation or with newly detected PCC/PG and changes in the management of index patients.

Data Sources and Search Strategies

A comprehensive search of several databases was conducted from inception to June 2012. These included Ovid MEDLINE In-Process and Other Non-Indexed Citations, Ovid MEDLINE, Ovid EMBASE, Ovid Cochrane Database of Systematic Reviews and Scopus. The search strategy was designed and conducted by an experienced librarian with input from the study's principal investigator. Controlled vocabulary supplemented with keywords was used to search for pheochromocytoma/paraganglioma, and genetic testing. The detailed strategy is available in the appendix of the systematic review. Experts in the field were contacted to confirm completeness of the search.

Study Selection

Two reviewers working independently screened titles and abstracts, excluding studies that were not germane to the research question or study design. Reviewers then conducted a second stage of screening, after obtaining full-text versions of articles. Chance-adjusted inter-rater agreement for this stage was high (kappa statistic = 0.98), and discrepancies were resolved by discussion (see Figure 1 in the systematic review document).

The Incremental Benefit of Functional Imaging in Pheochromocytoma/Paraganglioma; A Systematic Review

Study Eligibility

Reviewers searched for full publications in any language of studies with any design that included patients with biochemically proven PCC or PG who underwent anatomic imaging (AI) modalities (computed tomography [CT] or/and magnetic resonance imaging [MRI]) plus additional functional imaging (FI) for PCC/PG localization. The outcomes of interest were the added diagnostic benefit attributable to FI in localizing primary tumors or metastasis, change in surgical intervention, completeness of surgical resection, resolution of symptoms and further characterization of lesions. Reviewers also looked for descriptions of harm resulting from the use of FI. This review did not aim to compare the sensitivity or specificity of each imaging modality.

Data Sources and Search Strategies

A comprehensive search of several databases from each database's earliest inception to June 2012 was conducted. The databases included Ovid Medline In-Process and Other Non-Indexed Citations, Ovid MEDLINE, Ovid EMBASE, Ovid Cochrane Central Register of Controlled Trials, Ovid Cochrane Database of Systematic Reviews, and Scopus. The search strategy was designed and conducted by an experienced librarian with input from the study's principle investigator. Controlled vocabulary supplemented with keywords was used to search for pheochromocytoma/paraganglioma, CT scan/MRI, and functional imaging. The complete strategy is available in the appendix of the systematic review document.

Study Selection

Two reviewers performed first-stage screening of titles and abstracts based on the research question and study design. Retrieved articles underwent a second-stage screening in full text. Chance-adjusted inter-rater agreement for this stage was high (kappa statistic = 0.96). Discrepancies were resolved by discussion.

Number of Source Documents

Testing for Germline Mutations in Sporadic Pheochromocytoma/Paraganglioma: A Systematic Review

321 records were screened, 62 full-text articles were assessed for eligibility, and 31 studies were included in the qualitative synthesis.

The Incremental Benefit of Functional Imaging in Pheochromocytoma/Paraganglioma: A Systematic Review

306 records were screened, 88 full-text articles were assessed for eligibility, and 32 studies were included in the qualitative synthesis.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Quality of Evidence

+OOO Denotes very low quality evidence

++OO Denotes low quality evidence

+++O Denotes moderate quality evidence

++++ Denotes high quality evidence

Methods Used to Analyze the Evidence

Meta-Analysis

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

The Task Force reviewed primary evidence and commissioned two additional systematic reviews (see the "Availability of Companion Documents" field) to support the guideline.

Testing for Germline Mutations in Sporadic Pheochromocytoma/Paraganglioma: A Systematic Review

Data Extraction

Two reviewers, working independently, extracted the following data items from each included study: country of origin, study design, patient age and gender, the number of patients with sporadic tumors, and the number of germline mutations. When studies did not explicitly report their criteria for defining sporadic tumors (absence of family history of pheochromocytoma/paraganglioma [PCC/PG], syndromic features, bilateral disease and metastatic disease), authors were contacted to obtain this information, determine the number of patients that fulfilled all or most of these criteria, and their associated frequency of germline mutations. If reviewers were unable to obtain data from the authors, the population within each study that complied with the highest number of criteria was examined. It was also noted when studies included selected populations, such as patients who had had prior negative genetic analysis for germline mutations or patients already known not to have germline mutations (e.g., multiple endocrine neoplasia type 2 [MEN2], neurofibromatosis type 1 suppressor gene [NF1], von Hippel-Lindau disease tumor suppressor gene [VHL]). Reviewers also extracted information about the germline mutations, the location of the PG (head and neck vs thoraco-abdominal) and the impact of genetic testing on the number of new mutations or new tumors identified in family members of the index cases. For the index cases, it was noted whether there were changes in surgical management (Yes, No, not assessed [NA]) or localization of any previously unidentified lesions (Yes, No, NA). Finally, any harm or cost associated with genetic testing was also extracted. If data regarding the impact of genetic testing was not available in each study, reviewers attempted to contact authors for this information.

Quality Assessment

Two reviewers working independently assessed the methodological quality of included observational studies using the Newcastle-Ottawa Scale. This instrument assesses the protection against bias by evaluating how the investigators selected patients, evaluated outcomes and ensured the comparability of study groups.

Data Synthesis

Reviewers used descriptive statistics to present the results, whereby the proportions show the frequency of each mutation in patients tested. There were no comparative data showing the difference in outcomes (benefits or harms) between patients tested and patients who elected to forego testing.

The Incremental Benefit of Functional Imaging in Pheochromocytoma/Paraganglioma: A Systematic Review

Data Extraction

For each eligible study, two reviewers extracted the following data items: country in which the study was conducted; study design; number of patients followed; patient age and gender; type of functional imaging (FI) modality; whether the study included PCC, PG or both; and nature of the lesions (benign, malignant or both). Cases were classified as syndromic or sporadic depending on the presence or absence of family history of pheochromocytoma and paraganglioma, syndromic features and/or multiple or bilateral disease. Additionally, anatomic imaging (AI) was categorized as "localized" when the computed tomography/magnetic resonance imaging (CT/MRI) was confined only to the area of interest (e.g., the abdomen) and "non-localized" when the CT/MRI included at least the chest and abdomen. Reviewers defined the main outcomes in the following ways: i) when tumors were identified in the adrenal gland/s, this was considered as "primary localization of PCC"; when tumors were identified in the abdomen this was counted as "primary localization of PG;" ii) designation of "malignant lesions" required imaging findings consistent with local invasion or—in the case of retrospective studies—a surgical sample consistent with malignancy; iii) when lesions were detected distant from what was considered the primary location or in anatomic locations where chromaffin tissue is not otherwise found, or—in the case of retrospective studies—a surgical sample consistent with metastatic disease existed, they were considered "metastatic lesions;" iv) "change in surgical intervention" was used to describe those situations in which FI avoided an unnecessary surgical procedure, triggered an intervention or resulted in any change in the surgical approach (e.g., laparotomy instead of laparoscopy); v) if the postsurgical biochemical profile for PCC or PG reached normal limits, it was called "completeness of surgical resection;" vi) when postsurgical follow-up confirmed the absence of initial symptoms clearly related to PCC and PG, this was considered a "resolution of symptoms;" vii) when FI was able to clarify or further elucidate any feature of the lesion deemed to be important (i.e., vascularity, invasion to adjacent organs and size), this was counted as "further characterization of lesions." Finally, reviewers extracted any described harm associated with the utilization of FI (e.g., cost, side effects of contrast material or hospitalization days).

Quality Assessment

Two reviewers working independently assessed the methodological quality of included observational studies using items of the Newcastle-Ottawa Scale. This instrument evaluates the manner in which the investigators selected patients, measured the outcomes, and ensured the comparability of study groups.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Participants

The Task Force included a chair selected by the Endocrine Society Clinical Guidelines Subcommittee (CGS), seven experts in the field, and a methodologist. The authors received no corporate funding or remuneration.

Evidence

This evidence-based guideline was developed using the Grading of Recommendations, Assessment, Development, and Evaluation (GRADE) system to describe both the strength of recommendations and the quality of evidence. The Task Force reviewed primary evidence and commissioned two additional systematic reviews.

Consensus Process

One group meeting, several conference calls, and e-mail communications enabled consensus. Committees and members of the Endocrine Society, European Society of Endocrinology, and American Association for Clinical Chemistry reviewed drafts of the guidelines.

Rating Scheme for the Strength of the Recommendations

Strength of Recommendation

- 1 Indicates a strong recommendation and is associated with the phrase "The Task Force recommends."
- 2 Denotes a weak recommendation and is associated with the phrase "The Task Force suggests."

Cost Analysis

A cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

The Endocrine Society Clinical Guidelines Subcommittee and Clinical Affairs Core Committee provided careful critical review of earlier versions of this manuscript and helpful comments and suggestions. Members of the Endocrine Society kindly reviewed the draft version of this manuscript when it was posted on the Society's website and sent additional comments and suggestions.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Suspicion, confirmation, localization, treatment, and resection of pheochromocytoma and paraganglioma (PPGL) are important to decrease cardiovascular morbidity and mortality.
- Patients with PPGLs benefit from genetic counseling before and after germline mutation testing in order to be informed about the different suspected inherited diseases and their diagnosis and treatment, the diagnostic performance of corresponding genetic testing, and the familial risk of transmission.
- For familial disease, detection of a mutation in the proband may result in earlier diagnosis and treatment in other family members upon routine screening.
- Because some PPGLs have malignant potential, earlier detection and treatment may be important to prevent metastatic disease.
- See the "Values and Preferences" section in the original guideline document for an assessment of benefits and harms for each recommendation.

Potential Harms

- Clinicians need to be aware of both false-positive and false-negative diagnostic test results.
- Drawing blood in the seated position for measurement of plasma metanephrines entails an increased likelihood of false-positive results and a
 need for follow-up with sampling in the supine position.
- A misinterpretation of genetic testing or incorrect results can lead to deleterious consequences for the patient and his or her family.
- α-adrenergic receptor blockers used to minimize perioperative complications have the potential to cause side effects, such as hypotension and reactive tachycardia.
- The major potential postoperative complications are hypertension, hypotension, and rebound hypoglycemia. The recommendation that blood pressure, heart rate, and plasma glucose levels should be closely monitored for 24–48 hours is mainly based on retrospective studies and institutional experience. Because of potential adrenal insufficiency, particular attention needs to be paid to patients who undergo: 1) bilateral adrenalectomy; 2) bilateral cortical-sparing adrenalectomy; or 3) unilateral cortical-sparing adrenalectomy of a sole remaining adrenal gland. There are numerous case reports of postsurgical hypoglycemia but no studies documenting its exact prevalence.
- Mortality rate with laparoscopic adrenalectomy is about 1%, and the conversion rate and transfusion rate are about 5% (rate of conversion to open resection is influenced by tumor size and surgeon experience).
- Partial adrenalectomy inevitably leaves some adrenal medullary tissues with risk for recurrent pheochromocytoma. The resulting potential for
 reoperations with higher conversion and complication rates must be balanced against the risks associated with chronically treated adrenal
 cortical insufficiency.
- See the "Values and Preferences" section in the original guideline document for an assessment of benefits and harms for each recommendation.

Contraindications

Contraindications

- Use of ¹²³I-metaiodobenzylguanidine (MIBG) scintigraphy is contraindicated in pregnant women.
- The use of positron emission tomography (PET) imaging modalities is contraindicated in pregnant women.

Qualifying Statements

Qualifying Statements

- Clinical Practice Guidelines are developed to be of assistance to endocrinologists and other health care professionals by providing guidance
 and recommendations for particular areas of practice. The Guidelines should not be considered inclusive of all proper approaches or
 methods, or exclusive of others. The Guidelines cannot guarantee any specific outcome, nor do they establish a standard of care. The
 Guidelines are not intended to dictate the treatment of a particular patient. Treatment decisions must be made based on the independent
 judgment of health care providers and each patient's individual circumstances.
- The Endocrine Society makes no warranty, express or implied, regarding the Guidelines and specifically excludes any warranties of
 merchantability and fitness for a particular use or purpose. The Society shall not be liable for direct, indirect, special, incidental, or
 consequential damages related to the use of the information contained herein.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

Staff Training/Competency Material

For information about availability, see the Availability of Companion Documents and Patient Resources fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Lenders JW, Duh QY, Eisenhofer G, Gimenez-Roqueplo AP, Grebe SK, Murad MH, Naruse M, Pacak K, Young WF Jr, Endocrine Society. Pheochromocytoma and paraganglioma: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab. 2014 Jun;99(6):1915-42. [264 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2014 Jun

Guideline Developer(s)

The Endocrine Society - Professional Association

Source(s) of Funding

Funding for this guideline was derived solely from the Endocrine Society, and thus the Task Force received no funding or remuneration from commercial or other entities.

Guideline Committee

The Pheochromocytoma and Paraganglioma Guidelines Task Force

Composition of Group That Authored the Guideline

Task Force Members: Jacques W. M. Lenders (Chair); Quan-Yang Duh; Graeme Eisenhofer; Anne-Paule Gimenez-Roqueplo; Stefan K. G. Grebe; M. Hassan Murad; Mitsuhide Naruse; Karel Pacak; William F. Young Jr

Financial Disclosures/Conflicts of Interest

The Endocrine Society maintains a rigorous conflict-of-interest review process for the development of clinical practice guidelines. All Task Force members must declare any potential conflicts of interest, which are reviewed before the members are approved to serve on the Task Force and periodically during the development of the guideline. The conflict-of-interest forms are vetted by the Clinical Guidelines Subcommittee (CGS) before the members are approved by the Society's Council to participate on the guideline Task Force. Participants in the guideline development must include a majority of individuals without conflict of interest in the matter under study. Participants with conflicts of interest may participate in the development of the guideline, but they must have disclosed all conflicts. The CGS and the Task Force have reviewed all disclosures for this guideline and resolved or managed all identified conflicts of interest.

Conflicts of interest are defined by remuneration in any amount from the commercial interest(s) in the form of grants; research support; consulting fees; salary; ownership interest (e.g., stocks, stock options, or ownership interest excluding diversified mutual funds); honoraria or other payments for participation in speakers' bureaus, advisory boards, or boards of directors; or other financial benefits. Completed forms are available through the Endocrine Society office.

Financial Disclosures of the Task Force

Jacques W. M. Lenders, MD, PhD, FRCP (Chair)—Financial or Business/Organizational Interests: University of Dresden, Executive Committee member of PRESSOR (Pheochromocytoma and Paraganglioma Research Support Organization); Significant Financial Interest or Leadership Position: none declared.

Quan-Yang Duh, MD—Financial or Business/Organizational Interests: none declared; Significant Financial Interest or Leadership Position: none declared

Graeme Eisenhofer, PhD—Financial or Business/Organizational Interests: GWT (Gesellschaft für Wissens and Technologie), European Society of Endocrinology, ENSAT (European Network for the Study of Adrenal Tumors), PRESSOR, Eli Lilly; Significant Financial Interest or Leadership Position: none declared.

Anne-Paule Gimenez-Roqueplo, MD—Financial or Business/Organizational Interests: none declared; Significant Financial Interest or Leadership Position: none declared.

Stefan K. G. Grebe, MD, PhD—Financial or Business/Organizational Interests: none declared; Significant Financial Interest or Leadership Position: none declared.

M. Hassan Murad, MD*—Financial or Business/Organizational Interests: KER (Knowledge and Evaluation Research) Unit (Mayo Clinic); Significant Financial Interest or Leadership Position: none declared.

Mitsuhide Naruse, MD, PhD—Financial or Business/Organizational Interests: Japan Endocrine Society Council Member; Significant Financial Interest or Leadership Position: none declared.

Karel Pacak, MD, PhD, DSc—Financial or Business/Organizational Interests: none declared; Significant Financial Interest or Leadership Position: none declared.

William Young, Jr, MD, MSc—Financial or Business/Organizational Interests: The Endocrine Society; Significant Financial Interest or Leadership Position: none declared.

*Evidence-based reviews for this guideline were prepared under contract with the Endocrine Society.

Guideline Endorser(s)

American Association for Clinical Chemistry, Inc. - Professional Association

European Society of Endocrinology - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Guideline Availability

Electronic copies: Available from The Endocrine Society Web site	Electronic copies: Available from Th	e Endocrine Society Web site	
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Print copies: Available from The Endocrine Society, Phone: (301) 941.0210; Email: Societyservices@endo-society.org

Availability of Companion Documents

The following are available:

- Brito JP, Asi N, Gionfriddo MR, Norman C, Leppin AL, Zeballos-Palacios C, Undavalli C, Wang Z, Domecq JP, Prustsky G, Elraiyah TA, Prokop LJ, Montori VM, Murad MH. The incremental benefit of functional imaging in pheochromocytoma/paraganglioma: a systematic review [abstract]. Electronic copies: Available from The Endocrine Society Web site.
- Brito JP, Asi N, Bancos I, Gionfriddo MR, Zeballos-Palacios CL, Leppin AL, Undavalli C, Wang Z, Domecq JP, Prustsky G, Elraiyah
 TA, Prokop LJ, Montori VM, and Murad MH. Testing For germline mutations in sporadic pheochromocytoma/paraganglioma: a systematic
 review. Clinical Endocrinology; 2014 Jul 7 [Epub ahead of print]. 8 p. Electronic copies: Available to subscribers from the Clinical
 Endocrinology Web site

In addition, continuing medical education activities are available in the original guideline document

Print copies: Available from The Endocrine Society, Phone: (301) 941-0210; Email: Societyservices@endo-society.org.

Patient Resources

NGC Status

This NGC summary was completed by ECRI Institute on November 21, 2014. The information was verified by the guideline developer on December 4, 2014.

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